Diagnostic Clues to Frontal Fibrosing Alopecia in Patients of African Descent

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ABSTRACT

Importance: Frontal fibrosing alopecia has previously been reported as rare among patients of African descent. The authors present 18 cases of frontal fibrosing alopecia affecting African American patients and review all published cases of frontal fibrosing alopecia involving patients of African descent. **Observations:** Since 2010, there have been 66 published cases of frontal fibrosing alopecia among patients of African descent; 59 women, five men, and two cases of unknown gender. Frontal fibrosing alopecia is not uncommon among patients of African descent. In this study, the authors find that female African American patients may have fewer symptoms and unique clinical presentations. **Conclusion and relevance:** Frontal fibrosing alopecia is an entity that can be seen in patients with many different ethnic backgrounds, often with varying presentations. The diagnosis of frontal fibrosing alopecia must be considered in any patient of African descent who presents with frontotemporal alopecia. In the authors' patient population, there was a younger age of presentation. The presence of perifollicular hyperpigmentation along the hairline and concomitant facial hyperpigmentation may aid in making the diagnosis and distinguishing this entity from traction alopecia. (*J Clin Aesthet Dermatol.* 2016;9(4):45–51.)

Frontal fibrosing alopecia (FFA), a cicatricial alopecia, is currently accepted as a variant of lichen planopilaris (LPP) and is a challenging disorder to treat. Progressive, bilateral recession of the frontotemporal hairline is the defining feature and most consistent presentation of this disease. Kossard first described FFA in 1994 as a scarring hair loss generally seen in postmenopausal Caucasian women and resulting in progressive destruction and recession of the frontotemporal hairline.[1] Hair loss has also been reported to affect the eyebrows, occipital scalp, and the body. There are a rising number of cases seen in clinical practice, and the condition may be under diagnosed, particularly in the African American population. The differential diagnosis of frontotemporal hair loss includes traction alopecia, androgenetic alopecia, and alopecia areata, but FFA should always be considered in this population if marginal hair loss is present. Treatment is very challenging, and there have been no clinical trials to date to ensure efficacy or standardization of treatment regimens. This report of 18 cases will discuss the presentation, diagnosis, and treatment options for patients of African descent with FFA.

REPORT OF CASES

All patients were identified both clinically (based on evidence of frontotemporal hair loss) and microscopically (based on histological features of FFA). A retrospective chart review was then performed. Data collected from each chart can be seen in Table 1, including time course of presentation, clinical presentation, presenting symptoms, menopausal status, and treatment outcomes.

All 18 patients reviewed were African American females with the average age at diagnosis of 52 years (range 28–85 years of age)(Table 1). All patients had scalp biopsy results that conclusively demonstrated FFA. Seven out of 18 (39%) were premenopausal at the time of diagnosis and 7 of 18 (39%) were postmenopausal. Four had an unknown menstrual status. Seven patients (39%) related a family history of "hair loss." Three patients were lost to follow-up, but limited data available for these patients was included in the results.

Eighteen out of 18 (100%) demonstrated hair loss in the frontotemporal region. Eyebrow alopecia was clinically evident in 13 out of 17 (76%). Occipital hair loss was seen in

DISCLOSURE: The authors report no relevant conflicts of interest.

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TABLE 1. Patient demographics and clinical findings

PATIENT	FAMILY HISTORY	PRE/POST MENOPAUSE	HAIR WASHING FREQUENCY	GROOMING PRACTICES BEFORE DIAGNOSIS	GROOMING PRACTICES AFTER DIAGNOSIS	TREATMENT REGIMEN	CLINICAL STATUS AT TIME OF STUDY
1	Yes	Post	1x per week	None	Heat styling	Topical steroids, doxycycline	Stable
2	No	Post	1x every 2 weeks	Relaxer and heat styling	None	Topical steroids	Ongoing hair loss
3	No	Post	1x per 3 weeks	Relaxer and hair color	None	Antifungal shampoo	Ongoing hair loss
4	Yes	Post	1x per week	Relaxer and heat styling	None	Intralesional steroids	Stable
5	Yes	Post	every 1–2 weeks	None	None	Hydroxychloroquine	Stable
6	No	Unknown	1x per week	Relaxer and heat styling	Heat styling	Topical steroids, intralesional steroids, doxycycline, antifungal shampoo	Ongoing hair loss
7	No	Pre	1x per week	Relaxer	None	Topical steroids, intralesional steroids	Stable
8	Yes	Post	n/a	n/a	n/a	n/a	n/a
9	Yes	Pre	1x every 2 weeks	Relaxer	None	Topical steroids, intralesional steroids, doxycycline, antifungal shampoo	Stable
10	Yes	Unknown	n/a	n/a	n/a	Topical steroids, intralesional steroids, hydroxychloroquine	n/a
11	Yes	Pre	1 x per week	Relaxer and heat styling	None	Topical steroids, doxycycline	Stable
12	No	Unknown	1 x every 2 weeks	Heat styling	Heat styling	Topical steroids, antifungal shampoo	Hair regrowth at frontal scalp
13	No	Unknown	4 x per week	Relaxer	None	Topical steroids, antifungal shampoo	Ongoing hair loss
14	No	Pre	1 x per week	Relaxer	None	Minoxidil	Stable
15	No	Pre	1 x per week	Relaxer and heat styling	Heat styling	none	Ongoing hair loss
16	No	Pre	1 x per week	Relaxer	Relaxers	Topical steroids, doxycycline, hydroxychloroquine	Ongoing hair loss
17	No	Post	1 x every 1.5–2 weeks	Weaves	Heat styling	Topical steroids, doxycycline	Ongoing hair loss
18	No	Pre	every other day	Hair color	None	Topical steroids, doxycycline	Stable

Continued on the next page

TABLE 1 cont. Patient demographics and clinical findings

PATIENT	DURATION OF HAIR LOSS	AREAS INVOLVED	PIGMENTARY CHANGES	SCALP PAIN	PUSTULE/ PAPULE	PRURITUS	ERYTHEMA	FLAKING
1	2 years	Frontotemporal; eyebrows	No	no	no	yes	no	no
2	5 years	Frontotemporal	Yes, PIH on face	no	papules	yes	no	no
3	4 years	Frontotemporal; eyebrows; eyelashes; vertex; occiput	No	no	no	yes	no	no
4	2 years	Frontotemporal; eyebrows	No	no	no	yes	no	no
5	10 years	Frontotemporal; eyebrows	Yes, follicular hyperpigmentation	no	no	no	no	no
6	2.5 yrs	Frontotemporal; eyebrows	Yes, PIH on face	yes	no	yes	no	no
7	5 years scalp, 1 year brows	Frontotemporal; eyebrows	No	yes	yes	yes	no	no
8	n/a	Frontotemporal; Unable to confirm other areas	n/a	n/a	n/a	n/a	n/a	n/a
9	7 years	Frontotemporal; eyebrows	No	yes	no	yes	no	no
10	1 year scalp, 4 months brows	Frontotemporal; eyebrows	Yes, hypopigmentation of eyebrows	n/a	n/a	yes	no	no
11	1 year	Frontotemporal	No	no	no	yes	no	yes
12	1 year	Frontotemporal; eyebrows	No	no	no	no	yes	no
13	1 year	Frontotemporal; eyebrows; occiput	Yes, PIH on face	yes	no	no	no	no
14	3 years	Frontotemporal	No	no	no	yes	no	yes
15	3 years	Frontotemporal; eyebrows	Yes, follicular hyperpigmentation	no	no	no	no	no
16	2 years	Frontotemporal; eyebrows; occiput	Yes, follicular hyperpigmentation	no	no	no	no	no
17	7 years	Frontotemporal; eyebrows	No	no	no	yes	yes	yes
18	2 years	Frontotemporal	Yes, follicular hyperpigmentation	no	no	yes	yes	no

All women included in the case series self-classified as African American and all had a biopsy-proven diagnosis of FFA

TABLE 2. Published reports of FFA in patients of African descent

AUTHORS	# CASES	ETHNICITY	DESIGN	AVERAGE AGE	MENSTRUAL STATUS	OUTCOME
Chew et al 2010[11]	1 woman	Afro-Caribbean	Case series, 13 patients	42	Not reported	Not reported
Samrao et al 2010[5]	2 cases, gender not reported	African American	Retrospective chart review, 36 patients	Not reported	Not reported	Not reported
Donati et al 2011[13]	2 women	Black	Case series, 4 patients	One 53, one 56	1 postmenopausal, 1 perimenopausal	Not reported
Miteva et al 2012[3]	10 women, 1 man	Black	Retrospective multi-center review, 141 patients	53–72	Postmenopausal	Not reported
Ramaswamy P et al 2012[9]	1 man	African American	Case series, 3 patients	40	n/a	Not reported
Dlova et al 2013[14]	3 women	African	Case series, 10 patients	56	Not reported	Not reported
Dlova 2013[4]	21 women, 1 man	African	Retrospective chart review, 24 patients	42	13 pre, 7 post, 1 unknown	Not reported
Ladizinski B et al 2013[10]	1 woman	African American	Retrospective chart review, 19 patients	63	Postmenopausal	Not reported
Dlova NC and Goh CL 2015[15]	1 man	African	Case report, 1 patient	35	n/a	Stabilized
Dlova et al 2013[2]	17 women, 1 man	African	Retrospective chart review, 20 patients	42	13 pre, 5 post	Not reported
Vano-Galvan S et al 2014[6]	3 women	Black	Retrospective chart review, 355 patients	Not reported	Not reported	Not reported

4 out of 17 (23.5%) while vertex and eyelash involvement was only seen in one patient. In addition to frank hair loss, 12 out of 17 (70%) patients complained of itching and 4 out of 16 (25%) complained of scalp pain. Clinical examination showed changes in perifollicular pigment in 4 out of 17 (24%), with erythema and flaking each noted in three patients (18%). Patients did not commonly present with papules or pustules on the scalp, which can sometimes be found in other forms of cicatricial alopecia.

In regard to hair grooming practices, 15 out of 17 (88%) patients reported washing their hair once a week or less frequently. Eleven out of 16 (69%) used chemical relaxing agents prior to the diagnosis of FFA, and most of these patients discontinued use of relaxers after diagnosis. Only one patient continued to use relaxers after receiving a diagnosis of FFA. Six out of 16 (38%) used heat for hair styling prior to diagnosis of FFA and 5 out of 16 (31%) continued to heat style their hair after diagnosis. Use of hair color or hair weaves was uncommon.

Since no standardized treatment regimen exists for this disease, empiric treatments were employed based on patients' clinical presentation and reported symptoms. Topical corticosteroids were the most commonly used medications and were prescribed to 12 of 17 (71%) patients while 4 of 17 (24%) patients received intralesional corticosteroid injections. Antibiotics, most commonly doxycycline, were prescribed to 7 out of 17 (41%), 5 out of 17 (29%) patients were prescribed antifungal shampoos, and 3 of 17 (18%) were treated with hydroxychloroquine. Only 8 of 16 (50%) experienced stabilization of hair loss while 7 of 16 (44%) continued to have hair loss. One patient reported some hair regrowth in the frontotemporal region.

DISCUSSION

A PubMed search for "frontal fibrosing alopecia" revealed 99 total publications, 11 of which involved cases of FFA in patients of African descent. Between 2010 and 2013, there have been 66 reported cases of FFA in patients of African descent (Table 2). In many of these cases, there was associated traction alopecia.[2,3] Some cases were observed to

stabilize over time. There appears to be a rapidly escalating incidence of FFA with increasing reports of the disease occurring among Caucasian women and men and women of African, Hispanic, and Asian ethnicities.

The authors' data demonstrated that 39 percent of their patients presented before menopause and other studies involving a demographically similar cohort found 65 to 75 percent presented pre-menopause.[2,4] In contrast, studies involving Caucasian women have found 5 to 17 percent of patients present pre-menopause.[5] The average age of diagnosis was 52 in the authors' study while current literature shows a range from 55.5 to 63 in Caucasians and 40 to 42 in Africans.[2,4] The small sample size of this study limits the ability to determine the statistical significance as compared to previously published data; however, a younger age of presentation may be characteristic in this population.

FFA causes follicular scars with a progressive course that can have devastating effects on an individual's emotional status and appearance. The disease progresses slowly for several years until it stabilizes or remits spontaneously.[6] Some cases may also exhibit circumferential hair loss as seen in Figure 1A. Determining the precise age of onset is difficult because of the slowly progressive nature of the disease. Many individuals do not notice the hair loss until it becomes cosmetically evident after a significant amount of follicular destruction has already occurred.[7] It is possible that many cases are diagnosed post-menopause although the actual onset of disease may have occurred before menopause.[7] Since most cases of FFA are seen in Caucasians, it is important to consider this disease in patients of other racial and ethnic groups, including Africans,[2] Latinos,[6] Asians,[4,8] African Americans,[5,9,10] and all patients of African descent.[11] Symptoms, such as follicular erythema, scaling, and papules and pustules of the scalp, which commonly occur in other scarring alopecias, were not as frequently seen in this cohort of African American patients with FFA. Since these characteristics are generally seen in the early stages of disease, it is possible that patients presented after this stage of disease when those symptoms were no longer prominent. An alternate explanation includes the possibility that symptomatology is not as common in patients of African descent or is less apparent due to the increased pigmentation of the skin. Future research is needed to clarify this. Diagnosis in this patient population can be difficult if common presenting symptoms are absent. As with other forms of scarring alopecia, diagnosis is best ascertained through biopsy at the border of the alopecic zone where hair is still present.[7]

Dermoscopy can be used to assist in making the diagnosis of FFA. Dermoscopic findings include loss of orifices, perifollicular erythema, and scale.[8] Another interesting finding in the authors' patients was the presence of follicular hyperpigmentation, as seen in Figures 1A and 1B. This speckled appearance of the frontal hairline has not been described for FFA, and appears to be a unique characteristic more commonly seen in darker skinned patients. There is one case study in Brazil[12] where perifollicular hyperpigmentation was observed by dermoscopy in 2 out of 4 patients with lichen planopilaris and 1 out of 5 patients with discoid lupus. Eyebrow alopecia, which has been well reported in the literature for Caucasian patients, was also seen in 76 percent of African American patients in this study. Therefore, eyebrows

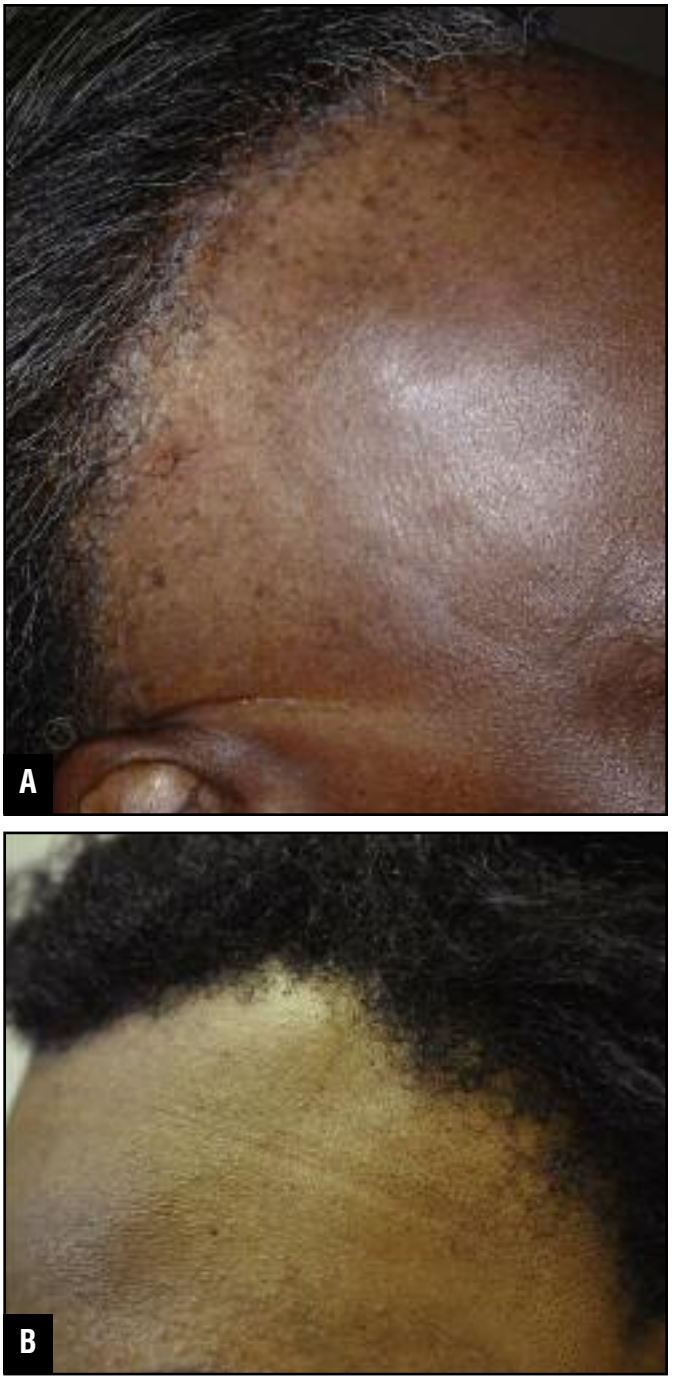

Figure 1. Clinical presentations of patients with FFA: A) Frontal fibrosing alopecia involving the hairline circumferentially in a middle aged African American female with pronounced perifollicular hyperpigmentation. B) A patient with frontal fibrosing alopecia demonstrating loss of hair from the frontal hairline that can sometimes clinically mimic traction alopecia.

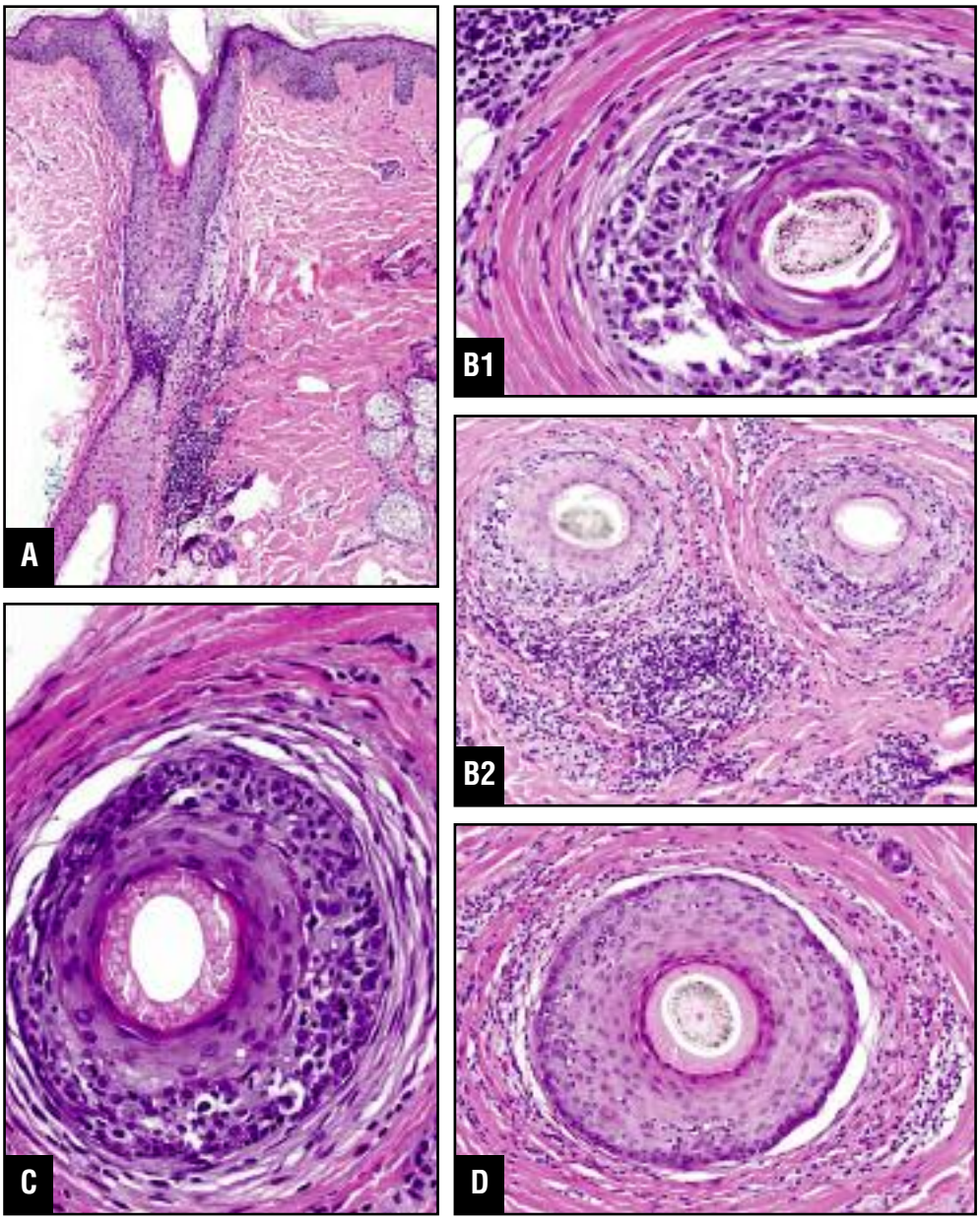

Figure 2. Histologic features of FFA: A) The inflammatory lymphocytic infiltrate involves the isthmus and infundibulum of this vertically sectioned follicle; B) There is damage to the basal and epibasal follicular epithelium, with blurring of the epithelial/stromal border, and apoptotic/dyskeratotic cells in the outer root sheath. The inner root sheath may be lost (B1) or intact (B2) in affected follicles; C) There is migration of lymphocytes into the basal and epibasal epithelium as well as prominent basilar alteration and concentric, lamellar fibroplasias; D) Artifactual clefting between epithelium and stroma is present; note the intact inner root sheath, which excludes central centrifugal cicatricial alopecia from the differential diagnosis.

in African patients with FFA.[4] Dlova described 24 cases of LPPigm which preceded FFA. These patients presented with hyperpigmentation in sun-exposed areas and the diagnosis was confirmed histologically. Facial hyperpigmentation was seen in the authors' patients; however, biopsy for LPPigm was not performed.

The histopathologic findings for the authors' patients with FFA are identical to those seen in LPP[3,7] and cannot be differentiated from those seen in Caucasians with FFA.[7] Biopsy specimens from the authors' patients showed many or all of the following features: an inflammatory lymphocytic infiltrate involving the isthmus and infundibulum of the hairs (Figure 2A), vacuolar interface alteration of basilar and epibasilar follicular epithelium (Figure 2B), migration of lymphocytes into the epithelium (Figure 2D), dyskeratotic cells in the outer root sheath (Figures 2B and 2C), concentric perifollicular fibroplasia (Figures 2B and 2C), prominent artifactual clefting between the epithelium and stroma (Figure 2D), and subsequent destruction of follicles. Some involved follicles have intact inner root sheaths (Figures 2B–2D), a feature that excludes central centrifugal cicatricial alopecia (CCCA) from the differential diagnosis. A preponderance of inflammation affected the terminal hairs, although an occasional vellus hair was inflamed.

Most authors stress the histological similarities between FFA and LPP. However, some authors have found that FFA specimens show less follicular inflammation and contain more apoptotic cells than in LPP,[7] but these features are variable from patient to patient and even between follicles in the same specimen. None of the authors' FFA specimens showed an inflammatory infiltrate involving the interfollicular epidermis; other authors have noted the same absence of interfollicular epidermal involvement in

should be carefully examined when evaluating African American patients with fronto-temporal hair loss. Alopecia of the upper limbs has also been reported in patients with FFA with biopsy revealing features of LPP, indicating that this disease can have a generalized presentation in many patients.[11]

Lichen planus pigmentosus (LPPigm) has been reported

Caucasian patients.[7] Direct immunofluorescence was not performed on any of the authors' specimens.

Treatment regimens are currently provider-dependent, as no clinical trials on effectiveness for FFA have been performed to establish a standard of care. For patients in this study, treatment was initiated upon diagnosis to help with symptomatic relief and to attempt to halt further progression of the disease. The following medications have been reported to be useful: hydroxychloroquine, doxycycline, and mycophenolate mofetil.[5]

CONCLUSION

There are some notable findings in this study that have not been reported in the literature and should be recognized in patients of African descent with frontotemporal hair loss. These findings could aid in clinical diagnosis of FFA and distinguish it from traction alopecia. Age of onset tends to be earlier according to the authors' sample population, but the significance of this age difference is not clear. Follicular hyperpigmentation in the areas of hair loss, the likely result of postinflammatory changes, is pronounced in a subset of these patients. Clinical findings of erythema and follicular papules, common in other scarring alopecias, were uncommon presenting characteristics.

Treatment options vary widely with a lack of clinical trials to determine efficacy of treatment. Increased research and trials for treatment regimens are needed to aid dermatologists in halting and reversing this potentially devastating disease process.

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